



Case Report

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A Case of Successful Cardiopulmonary Resuscitation in a Pregnant Woman with Eisenmenger's Syndrome



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Abstract

Patients with Eisenmenger's syndrome are advised to avoid pregnancy due to high maternal and intrauterine mortality. Mortality during pregnancy in patients with Eisenmenger's syndrome reaches 30 - 50%. A 37-year-old pregnant patient, at a gestational age of 14/15 weeks, was admitted to the Almazov National Medical Research Center for abortion due to pulmonary arterial hypertension and Eisenmenger's syndrome. Due to the In view of the complete presentation of the chorion, it was planned to perform curettage of the uterine cavity after temporary embolization of the uterine arteries. After introduction of antibacterial drug in the perioperative period, acute decompensation of heart failure, hypotension and the decrease in Ejection Fraction (EF) by transthoracic echocardiography (TTE) were observed, which required cardiopulmonary resuscitation and ECMO extra-corporeal membrane oxygenation (ECMO) implantation. This condition was assessed regarded as the anaphylactic shock. It was possible to stabilize the patient after switching to ECMO was successful. A spontaneous miscarriage has occurred in the intensive care unit. On the next day, vasopressor and inotropic support was discontinued, the patient was extubated and the ECMO system was explanted on the 5th day. Subsequently, Pulmonary Arterial Hypertension (PAH). PAH-specific therapy was expanded, the patient was activated. On the 29-th day after the termination of pregnancy the patient was discharged with recommendations for a consultation at the transplant center. Few months later the patient rReadmitted to the Almazov Center a for a vaginal hysterectomy.

Keywords: Eisenmenger's syndrome Eisenmenger syndrome; Anaphylactic shock; Extracorporeal membrane oxygenation; Cardiopulmonary resuscitation; Pulmonary hypertension; Pregnancy

Background

The beginning of the 21-st century is marked by a steady upward trend in the number of adult patients the congenital heart disease (CHD). In developed countries, the number of adult patients with CHD has began to exceed the number of children with congenital heart defects [1]. This situation, of course, reflects the success of cardiac surgery and cardiology, the possibility of radical correction of congenital heart defects at an early age with a complete restoration of the functional state of patients [2]. However, in some patients, surgical interventions are only palliative in nature, and in some cases severe pulmonary hypertension persists. Unfortunately, there is still a number of patients who did not managed with proper cardiac surgical care in childhood and subsequent development of severe hemodynamic

disorders later in their lives, up to the formation of Eisenmenger's syndrome.

Eisenmenger's syndrome is marked by the development of a shunt of blood from the right side to the left side of the heart, resulting in defects of systemic and pulmonary circulation, which occur due to the formation of severe pulmonary hypertension. The excess pressure in the pulmonary circulation is higher than the pressure in the systemic circle. the Eisenmenger's syndrome develops with prolonged presence of uncorrected defects of the interventricular and interatrial septa, patent ductus arteriosus, tetralogy of Fallot, etc.

Patients with Eisenmenger's syndrome are characterized by:

- a) Cyanosis.
- b) Significant decrease of the oxygen content in the arterial blood.
- c) Large, rounded fingernails or toenails.
- d) Hemoptysis (rather late symptom).
- e) Secondary polycythemia (high level of hemoglobin due to a compensatory mechanism)
- f) Heart failure III - IV according to New York Heart Association (NYHA) Functional Classification. NYHA
- g) Patients with Eisenmenger's syndrome are advised to avoid pregnancy due to high maternal and intrauterine mortality [3]. Mortality during pregnancy in patients with Eisenmenger's syndrome reaches 30 - 50% [4].

In accordance with current international clinical guidelines [3,4], pregnant women with Eisenmenger's syndrome should be treated in the following way:

- a) During pregnancy women with Eisenmenger's syndrome should be observed by a multidisciplinary team, including a cardiologist, an obstetrician-gynecologist and an anesthesiologist.
- b) During pregnancy, specific therapy for pulmonary arterial hypertension (specific therapy for PAH) should be evaluated and possibly modified.
- c) Patients with Eisenmenger's syndrome should be transferred to reference centers.
- d) Operative abdominal delivery is recommended.
- e) During the operative abdominal delivery, preference should be given to regional methods of anesthesia. It is advised to avoid intubation and mechanical ventilation.
- f) The intensive care for treatment of acute right ventricular failure must be available.

In the respective literature, a number of clinical cases of cardiopulmonary resuscitation (CPR) in patients with Eisenmenger's syndrome [5-8] are described. However, only one publication demonstrates successful case of CPR in a pregnant woman with Eisenmenger's syndrome [9]. The reason for the low effectiveness of CPR is associated with the pathophysiological features of Eisenmenger's syndrome. During indirect (as well as direct) heart massage, venous blood from the right sections enters the left sections of the heart, following into the systemic circulation. Extremely low lung perfusion during CPR is associated with extremely high pulmonary resistance, often exceeding that of the systemic circulation. Extremely low lung perfusion during CPR is associated with extremely high pulmonary resistance, often exceeding that of the systemic circulation. These circumstances make maintenance of the adequate arterial oxygenation during CPR in patients with Eisenmenger's syndrome hardly possible [10].

Clinical Case

Patient M., 37 years old, pregnant, at a gestational age of 14/15 weeks, was admitted to the Department of Pathology of Pregnancy on June 27th of, 2022 for abortion due to Eisenmenger's syndrome. The patient is stratified in class IV according to World Health Organization (WHO) - prolongation of pregnancy is associated with an extremely high risk of developing CVD and maternal mortality.

Preoperative Management

From the anamnesis: patient is aware of her heart disease - ventricular septal defect (VSD) - since the age of 14. Patient has been repeatedly examined in cardiac surgery clinics, with surgical treatment denied due to high pulmonary hypertension. In 2016, an abortion was performed by curettage of the uterine cavity. PAH - specific therapy was initiated (bosentan was taken irregularly, and patient didn't switch to sildenafil therapy).

Allergies anamnesis: notes an allergy on Actovegin (urticaria). All antibacterial drugs that she received prior to her current hospitalization were well tolerated, without any anaphylactoid reactions. Additional evaluation at the Almazov Center included a physical examination, transthoracic echocardiography (TTE), 24-hour electrocardiography (ECG) monitoring and a panel of laboratory tests. Patient's weight was 52kg, height was 149cm. Blood pressure (BP) was the same on the right and left arms - 120/70mm Hg. SpO₂ was 78-81% at rest on atmospheric air, SpO₂ was 74-75% with a light physical activity on atmospheric air, SpO₂ was 90% with oxygen inhalation. There were no signs of congestion in the systemic circulation.

The main TTE parameters are listed below. Aorta - 32mm; Left atrium - 35mm; Atrial septum has not been changed. End-diastolic volume of the left ventricle (EDVLV) - 51ml; end-systolic volume of the left ventricle (ESVLV) - 21ml; stroke volume - 30ml (SV); ejection fraction of the left ventricle (EF) (Simpson) - 59%. Right ventricular (RV) (4-chamber position) - 32mm; Right ventricular front wall (RVFW) - 10mm; TAPSE - 1.8cm; Right atrium (RA) - 41\42mm. Pulmonary artery (PA) - 24mm; Pulmonary artery systolic pressure (PAPsys) - 139mm Hg. Vena cava inferior - 20mm, inspiratory collapse > 50%. There were no significant valvular disorders. D - deformation of the interventricular septum (IVS). There was a defect in the membranous part of the IVS up to 13mm with a right to left shunt.

24-hour ECG monitoring: sinus rhythm.

The results of main biochemical tests are listed below. ALT - 13 U/l, AST - 22 U/l, bilirubin - 11µmol/l, creatinine - 36µmol/l, lactate - 1.0mmol/l, hemoglobin - 133g/l. NT-proB-type natriuretic peptide - 99.52 pg/ml.

According to the ultrasound of the pelvic organs the chorion has been detected on the anterior wall. The edge of the chorion was in the region of the internal os extended to 1/3 of the cervix.

Fetus was in transverse presentation.

Preoperative PAH-specific therapy was adjusted by the decision of the multidisciplinary team: it was recommended to take sSildenafil 20mg TID 3 times a day with further resumption of bBosentan 125mg BID 2 times a day after the termination of pregnancy.

Surgical termination of pregnancy was recommended due to full chorion presentation. Additionally, temporary embolization of the uterine arteries was recommended in order to minimize blood loss. Epidural anesthesia with invasive monitoring, including catheterization of the radial artery and internal jugular vein, has been chosen. In the Almazov center, all patients with large septal defects are recommended for IV administration of any medications through a built-in air trap (Sterifix 0.2µm, Luer Lock, low protei binding, B Braun, Germany) to prevent air embolism.

Surgical Procedure, Anesthesia, Cardiopulmonary Resuscitation

06/30/2022 09:40 - 10:20. The patient was prepared to surgical procedure in the cath-lab according to standard methods. 10:25. Antibacterial prophylaxis with aAmpicillin + sSulbactam, (Sultasin, Sintez, Russia) 3000 mg was initiated. However, during infusion of 1000 mg of antibiotics, the patient's condition worsened (nausea, vomiting, agitation, hypersalivation, loss of consciousness, hemodynamic depression up to 60/40 mm Hg, tachycardia up to 145 beats/min, SpO2 decreased to 56%), diffuse cyanosis.

10:25 - 10:45. Intensive therapy was started to ensure airway patency (oro-tracheal intubation and controlled mechanical ventilation (CMV) with FiO2 100%) and hemodynamic support including chest compressions. As anaphylaxis was suspected, Epinephrine bolus 100µg was given IV, further followed by it's titration 0.1 - 0.3µg/kg/min, Norepinephrine 0.1 - 0.5µg/kg /min was titrated for vasopressor support. Infus Non therapy and correction of acid-base balance were carried out, methylprednisolone 120mg was administered. To reduce pulmonary vascular resistance and improve the hemodynamic profile, nitric oxide was inhaled at a dose of 60 ppm (synthesis

from atmospheric air with the "Tianox" device, Russia). In the cath-lab operating room TTE and chest x-ray were performed, left ventricular injury, hemopericardium, pneumothorax and hemothorax were excluded.

10:45. Patient displayed supraventricular tachycardia up to 150 beats / min, arterial hypotension - 50/40-40/20mm Hg, central venous pressure (CVP) - 6mm Hg. Due to the lack of effectiveness of cardiopulmonary resuscitation, the team decided to start veno-arterial ECMO.

ECMO Procedure

10:45-11:30. Venous and arterial cannula implantations were performed, and an ECMO circuit was prepared.

11:30. ECMO-flow 3 l/min was initiated. It allowed to achieve stabilization of hemodynamics, an increase SaO2 up to 98%.

13:00. The patient was transferred to the adult Intensive Care Unit (ICU) on ECMO and mechanical ventilation.

15:15. Spontaneous miscarriage occurred while the patient was in the ICU. The patient received Carbetocin 100mcg to prevent bleeding. No bleeding from the genital tract was observed. The infusion of unfractionated Heparinum (UFH) required for ECMO was not carried out.

07/01/2022 00:15. Infusion of UFH 500 U/h was started. The patient underwent a course of antibiotic prophylaxis: Vancomycin 1gr BID i/v, Clindamycin 300mg TID i/v. Long-term vasopressor support (Norepinephrine) with further dose reduction.

05:30. ECMO flow was reduced to 2.5l/min against the background of satisfactory hemodynamic parameters and the cessation of vasopressor and support.

10:00. Patient displayed clear consciousness and muscle tone restoration, and was extubated (Table 1).

14:00. Patient was transferred to cardiac surgical ICU.

07/01/22-07/04/2022. The PAH therapy was adjusted, with increase to 4-component treatment: sildenafil (20mg TID); bosentan (125mg BID); selexipag (was started at dose 200µg BID); NO inhalation at dose 40ppm (Table 1&2).

Table 1: Acid-base gas exchange parameters.

Parameter	Before ECMO (O ₂ inhalation 8 l/min)	During CPR (CMV, FiO ₂ 100%)	After ECMO implantation (ECMO 3L/min)	After extubation (ECMO 2.2 L/min)	After ECMO explanation (O ₂ inhalation 8 l/min)
pH (a)	7,42	7,05	7,38	7,5	7,42
ΔpCO ₂	4,7	1,3	11,5	5,3	5,9
SpO ₂ (a)	94,0%	45,5	100	89	84,7
SpO ₂ (v)	63,5%	31,3	66,5	67,1	56,8
Lac (a)	1,0	9,8	11,2	0,9	0,9
Lac (v)	0,9	9,8	10,7	1,2	0,9
Hb g/l	139	132	130	114	138

Table 2: Parameters of systemic hemodynamics (calculation by the Fick method).

Parameter	Before ECMO	After ECMO explantation
Cardiac output, l / min	2,72	2,8
Cardiac index l/min/m ²	1,85	1,91
Systemic Vascular Resistance dynes*sec*cm ⁻⁵	2059	1914
Stroke volume, ml	33,97	30,8

07/08/2022. Inhalation of nitric oxide using «Tianox» device is turned off. The physical activation and rehabilitation program has been upgraded.

07/10/2022. The oxygen flow through the face mask was reduced to 2 l/min, SpO₂ – 87. When the patient breathed atmospheric air, SpO₂ decreased to 70 -75%.

07/11/2022. The patient was discharged from the cardiac surgical ICU and transferred to the cardiology department.

07/29/2022 The patient was hemodynamically and clinically stable against the background of three-component PAH-specific therapy. This therapy included Sildenafil (20mg TID), Ambrisentan (10mg OID), Bosentan (125mg BID), Selexipag (was titrated to the maximum tolerated dose - 1600µg BID The patient was discharged from the Almazov center with a moderate decrease in physical status. The multidisciplinary team recommended that the patient be admitted to the Shumakov Federal Research Center of Transplantology (Moscow, Russia) for a listing for transplantation of the heart and lung complex.

Follow Up After a Cardiopulmonary Resuscitation

It is important to note that our management of this patient was not limited to the period of early follow-up after successful cardiopulmonary resuscitation. From 11/01/22 to 11/14/22 the patient was hospitalized at the Shumakov Federal Research Center of Transplantology, where additional assessment was performed and decision about inclusion into the waiting list for cardiopulmonary complex has been made. The patient was examined by gynecologist. During the ultrasound scan of the pelvic organs a placental polyp has been found. The surgical treatment at Almazov center was recommended.

11/22/22 patient was admitted at the Almazov center to the gynecological department. Given the history of anaphylactic shock, the patient was consulted by an allergist, and several allergy tests and including tests with basophil activation were performed. The panel of medications for perioperative period has been examined.

A duplex scan of the arteries of the lower extremities was performed, stenosis of the right common femoral artery up to 30% was detected, the blood flow in the left common femoral artery was not disturbed. The surgical treatment plan was a vaginal hysterectomy. Epidural anesthesia with invasive monitoring, including catheterization of the radial artery and internal jugular

vein, was chosen.

Intra- and postoperative hemodynamics remained stable, vasopressor support was not required: BP - 120/60 - 130/65mm Hg, CVP +3mm Hg, sinus rhythm 60 -80 per minute, SaO₂ - 95% - 90% (oxygen inhalation at a flow rate of 6 l/min). The patient was transferred to the cardiology department on the 6th day and discharged from the Almazov center on the 28th day after the operation.

Discussion

At the Almazov center the management of pregnant women with Eisenmenger's syndrome is carried out by a multidisciplinary team-obstetricians-gynecologists, cardiologists, anesthesiologists and ICU physicians. WHO class IV patients are offered abortion for medical reasons. In view of the complete presentation of the chorion in the patient and the high risk of hemorrhagic complications, it was decided to perform temporary endovascular embolization of the uterine arteries, followed by curettage of the uterine cavity. under epidural anesthesia.

With the presence of a large ventricular septal defect and the presence of right-to-left shunting, CPR without ECMO would be ineffective, so a decision was made to implant in ECMO, after which the patient's condition stabilized.

We found only one publication about case of successful CPR in a pregnant woman with Eisenmenger's syndrome [9]. In this case, the ECMO system was available, but was not installed, since the cardiac arrest occurred due to ventricular fibrillation, which was stopped by defibrillation. Thus, a novel clinical case is suggested, confirming the effectiveness of ECMO in cardiopulmonary resuscitation in patients with Eisenmenger's syndrome.

Conclusion

Currently, the use of the ECMO system proves to be successful in CPR in patients with extremely severe disorders of intracardiac hemodynamics.

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